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INTRODUCTION

Although rickets, a disease related to vitamin D, is known to date back to Neanderthal man, an associated hearing loss was not reported until recently. Stamp and Baker (1976) investigated a family with recessive hypophosphatemic rickets, and found sensorineural hearing loss. Weir (1977) reported on this family and attributed this hearing loss to narrowing of the internal auditory canals, as identified radiologically.

Brookes (1983) identified sensorineural hearing loss in patients with vitamin D deficiency. He diagnosed the loss as cochlear, based on a variety of audiologic findings including loudness recruitment, small cochlear microphonics suggesting hair cell damage, and cochlear demineralization seen in petrous temporal bone polytomography. Amoung his ten subjects he also diagnosed Meniere's syndrome, otosclerosis, and inactive chronic suppurative otitis media.

In February 1984, an article appeared in the Annals of Internal Medicine, reporting an association between hearing loss and X-linked dominant hypophosphatemic bone disease, one of the many forms of rickets. In this article Davies et al. reported hearing loss in 19 out of 25 patients. These losses were found to be of

cochlear origin based on stapedius reflex thresholds, speech discrimination scores, tone decay, and loudness recruitment.

If it is true that defective bone mineralization can result in a loss of hearing sensitivity, this knowledge would be a step toward prevention or early rehabilitation. Knowledge of the location of the site of lesion and development of the hearing loss would be critical in identifying the cause and formulating prevention.

BACKGROUND

Rickets and osteomalacia both refer to the condition where the bones of the body fail to mineralize properly, resulting in a deformity of the long bones, as in bowed legs or knocked knees.

Normal bone is dense and strong. It is similar to fiberglass in that it is made of fibrous matrix (collagen) which is bound together with a glass-like substance made from minerals (hydroxyapatite). Two thirds of bone weight is the minerals phase.

As children grow, the normal skeletal system develops in three ways.

1. Growth--At either end of a long bone is a growth plate, made of cartilage. Within this growth plate, rapid cell growth occurs which enables a bone to lengthen. Subsequently, the cartilage ossifies, adding to the length of the long bone.

2. Modeling--Long bones are made up of a spongy center (trabecular bone), which is covered by a much more dense outer shell (compact bone), which grows in circumference similar to the growth rings of a tree. As new cells and new mineral are added to the outside of this compact bone, cells from the inner edge of the dense portion resorb, so as to create a hollow center for blood marrow.

3. Remodeling--Throughout the body, bone is continuously being regenerated to adjust to mechanical and metabolic needs. Also, if an injury occurs, the remodeling system rebuilds the damaged area (repair).

In rickets, the defective mineralization of bone affects all three types of bone development. Rickets can be definitively identified in children with an X-ray of the growth plate. (See figure 1.) During puberty, when growth is completed, the growth plates themselves become ossified, and are no longer indentifiable. Also, modeling of the bone ceases after the growth period. Therefore, the adult form of rickets, "osteomalacia", includes only the remodeling process. It can positively be identified and quantified by determining the amount of excessive unmineralized bone in a bone biopsy. (See Figure 2.)

Thus, rickets is a childhood disorder, which affects especially the growth plates of long bones while osteomalacia (the adult form) is observed at the skeletal remodeling sites. However,

they are both the result of disordered skeletal mineralization.

During childhood, interuption of bone lengthening results in short stature. Modeling defects show up as irregularities of the long bones, such as bowed legs or knocked knees. (See Figure 3.)

Remodeling defects in children or adults, are seen as irregular bone structure and mineral imbalance. Results of these may include ossification of liagments and joints, compression of the spinal column, premature closure of the skull structures, or dental abnormalities.

Current treatment for X-Linked Hypophosphatemic Rickets includes Calcitriol (a metabolite of vitamin D3), along with frequent doses of phosphate. Since this treatment will correct the growth, modeling, and remodeling defects normally seen in untreated patients, therapy should begin as early as possible. During treatment urine is monitored regularly to guard against vitamin D toxicity.

Mineralization of bone is dependent upon a very complex relationship between vitamin D, phosphate, and calcium. While diet provides some portion of the daily requirement for each of these, the entire required amount cannot be maintained by diet alone. In humans, the kidney must reclaim a large portion of phosphate, thus maintaining a balance in the body. (See Figure 4.)

Rickets and Osteomalacia may be caused by a lack of sunlight (5 to 20 minutes, a few times a week), or a dietary deficiency, resulting in a shortage of vitamin D, phosphate, or calcium. This form of rickets is almost unseen in the U.S. today, due to the fortification of milk with vitamin D and adequate nutrition.

However, there are other causes of rickets, including disorders of the kidney which prevent the reabsorption of necessary minerals into the blood system. Included in this category is X-linked hypophosphatemic rickets, the most common type of rickets in the United States. This cause of rickets can be identified with a blood test which shows normal calcuim levels, but a low blood phosphate level (due to renal wasting of phosphate). This low phosphate level prevents normal mineralization of the new bone growth.

X-linked hypophosphatemic rickets is a specific hereditary disorder passed along on the X chromosone as a dominant trait. Since males have only one X chromosone, they are more severely affected. A female has two X chromosomes, one involved and one unaffected, so she may show either a severe or a mild case.

Affected family members are quite easy to identify, but frequently a case occurs with no history of family involvment. These spontaneous occurrences are thought to be fresh mutations. The mutation will be transmitted to future generations on the

X-linked chromosome.

While audiologic findings in generalized skeletal disorders are rarely reported, a group of patients who had X-linked dominant hypophosphatemic osteomalacia was examined audiologically in the study mentioned earlier, by Davies et al. in England. Of twenty-five patients tested, hearing loss was identified in nineteen subjects (76%). Hearing loss was defined as a loss of hearing sensitivity that was greater than 15 dB in at least one frequency. Sixteen subjects showed a sensorineural hearing loss and the other three showed mixed losses (both a conductive and a sensorineural component to the loss). Davies et al. speculated that the loss was attributed to calcification of ligaments and bony overgrowth, reducing the movement of the basilar membrane and hair cell displacement.

Their subjects ranged from 11 to 75 years of age. Only four subjects were under the age of 20, and two of them were among the sample with normal hearing sensitivity. This suggests that age may have been a factor in the study from England.

Since this was the first report of an association between X-linked dominant hypophosphatemic rickets and hearing loss, the study was replicated, testing a similar population from Shriner's Hospital for Crippled Children in St. Louis. Because of the possible age effect, the study was broadened to include more children.

METHOD

SUBJECTS

Subjects included 19 patients with X-linked hypophosphatemic rickets or osteomalacia. This population included 11 children and 8 adults, who were relatives of the children. All of the children and one adult (patient #14) were being treated by Dr. Michael Whyte at Shriner's Hospital for Crippled Children in St. Louis, Missouri. Treatment of these patients for the bone disease included dosages of rocaltrol (a form of calcitriol) and K-phos (a phosphate diatary supplement). The possibility of ototoxic effects from these drugs is unknown, but unlikely.

Three patients (#13, 15, and 19) have never been treated for their bone disease. Patient #17 was given cod liver oil. Patients #4, 14, 16, and 18 have been treated with high dosages of vitamin D. Patient #12 was treated with Dristol. Patient #14 has also been treated with inderal and synthroid for episodes of vertigo over the past nine years. Patient #17 has been treated periodically with antivert to control attacks of vertigo. For two years patient #19 has taken various medications for a heart condition. None of these treatments are known to be ototoxic.

INSTRUMENTATION

Audiologic evaluations were conducted at Central Institute for

the Deaf in St. Louis, Missouri. All of the tests except the admittance measures were administered in sound treated booths, which were within the ANSI noise level specifications for audiologic examinations. The Grason-Stadler 1701 and 1715 audiometers were used to present pure tone, speech, and other test stimuli. All audiometric testing was conducted under the TDH 49 or TDH 50P earphones or Radioear B 70A or B 71 bone vibrators. Admittance measures were administered with the Grason-Stadler 1720 otoadmittance meter or the Grason-Stadler 27 immittance screener. All test equipment was calibrated to ANSI standards prior to testing.

The purpose of the test battery was to assess the sensitivity of the peripheral auditory nervous system and predict site-of-lesion if a hearing loss was discovered. Each ear was evaluated separately. The following tests were administered.

1. Air conduction thresholds for pure tones at octave intervals between 250 and 8000 Hz and bone conduction thresholds between 250 and 4000 Hz were obtyained with an ascending method. Hearing was not assessed below 0 dB HL. This testing was used to estimate hearing senditivity. Hearing sensitivity was classified as shown in Table 1.

Table 1 Levels of hearing loss

0-20 dB HL	Normal hearing
21-40 dB HL	Mild loss

41-55 dB HL Moderate loss

56-70 dB HL Moderate-Severe loss

71-90 dB HL Severe loss

91 or greater, Profound loss

2. Spondee thresholds or speech reception thresholds (SRT) were obtained with monitored live voice presentations using adult spondee word lists for patients age ten years and older. The children's spondee word lists were used with younger patients. This test was administered to examine the reliability of pure tone testing and determine the reference level for presentation of the speech discrimination test. SRT scores are generally consistent with the three frequency (500, 1000, and 2000 Hz) pure tone average (PTA). The SRT usually does not differ by more than 10 dB from the PTA.

3. Speech discrimination testing was administered at 40 dB re SRT. Monitored live voice versions of the CID W-22 test were used with patients 10 years of age and older. The Phonetically Balanced Kindergarten Word Test (PBK-50) was utilized with the younger population. Scores from these tests can help determine type of hearing loss.

4. Tone decay was measured at 500 Hz and 4000 Hz with the Olsen-Noffsinger method (1974).

5. Acoustic admittance measures were administered to evaluate

the acoustic characteristics of the tympanic membrane and estimate middle ear pressure. Results were obtained with a 220 Hz. probe tone and air pressure changes between -200 and +200 mm H2O. This testing was used to identify conductive abnormalities in the middle or external ear.

6. Acoustic Reflex Thresholds (ART), a measure of stapedius muscle reflexes, were assessed at 500, 1000, 2000, and 4000 Hz. These test results supply information concerning conductive and sensorineural hearing losses, as well as information about lesions in the VII or VIII cranial nerves, and lower brainstem.

7. The Weber test was administered at 500 Hz at 50 dB re threshold. This test was used to identify the ear with the greatest conductive component or the better bone conduction.

8. The Bing test was administered at 500 Hz at 50 dB re threshold. This test was included to help determine the presence or absence of a conductive hearing impairment.

RESULTS

The results of each test administered to each of the 19 patients in this study are shown in Table 2. The table is organized by age. The data show that 11 patients (58% of the sample) had no abnormal test results on any of the tests administered. Four patients (21% of the sample) had no measurable hearing loss, but exhibited some abnormal but insignificant test results (patients

#1, 2, 6, and 9). Each will be discussed separately. Therefore, a total of 15 out of the 19 patients (79% of the sample) had normal hearing sensitivity.

Patient #1 was 16 months old, and therefore, too young for most of the test battery. Results were obtained with Visual Response Audiometry. Responses to pure tone stimuli could only be obtained at 1000 and 2000 Hz and were 10 dB above normal in both ears. Speech awareness thresholds (SAT) were found to be well within the normal limits and acoustic admittance measures suggested normal middle ear functioning. Considering the patient's age, along with the test findings that were within the normal range, a hearing loss is unlikely. Some young children do not respond well to unfamiliar acoustic stimuli, such as pure tones. It is likely then that this child has normal hearing sensitivity. The results from this patient were not included in the study comparison because frequency specific information was was not obtained at the frequencies compared.

Test results for all but one test administered to patient #2 were within normal limits with no differences across ears. The results from the Weber test were inconsistent with the other test findings. This patient was only four years old and had to be reinstructed for the Weber test twice. It is possible that he did not fully understand the task. In view of the other findings it is likely that this child has normal auditory function.

Although no other abnormal auditory test findings were indicated by the audiologic test battery, admittance measures from patient #6 showed abnormally negative middle-ear pressure in the left ear. Patient #6 failed a hearing screening at her school two months prior to this evaluation. Since the present test results showed hearing sensitivity to be within the normal range and a slightly abnormal tympanogram, it is possible that patient #6 may have experienced middle ear problems that affected her hearing previously.

Results from patient #9 showed normal hearing sensitivity, normal speech discrimination scores, and no significant tone decay in either ear. However, acoustic reflex thresholds from this patient were elevated unilaterally and were not commensurate with the other test results. Since this patient did not have a history of tinnitus or vertigo, and exhibited normal test results on the majority of the tests, a retrocochlear lesion was not suspected. During the Weber test, patient #9 reported that the signal lateralized to her left ear. This result is also inconsistant with the other test findings. Results showed that patient #9 had identical hearing sensitivity across ears with no air-bone gap.

Since most audiologic tests require voluntary effort on the part of the patient, and therefore are not 100% reliable, the best interpretation of hearing is based on a battery of tests. The

results, that are consistent within the test battery for each of the 19 patients in this study, showed that 15 patients did have normal hearing sensitivity and no significant abnormal audiologic test findings.

That leaves four patients or 21% of the sample with measurable hearing loss. Three of those four were adults with sensorineural hearing loss (Patients #17, 18, and 19). Audiogram adjustments for age were not required. The fourth patient (#8) was a child with a conductive hearing loss. Hence, only four of the 19 patients in this study showed a measurable hearing loss. The audiologic results of these four patients will be discussed separately.

Test results from the child (patient #8) showed a mild unilateral conductive hearing loss with air conduction thresholds between 20 and 35 dB HL. Admittance measures revealed abnormally high static immittance (in equivalent volume units) bilaterally (right ear: 2.5 ml and left ear 3.9 ml) at 220 Hz. In addition, there was negative pressure in the ear with the hearing loss (left ear -150 da Pa). Lateralization of the signal to the left ear during the Weber test was consistant with the conductive hearing impairment in that ear. Results of patient #8 indicated that the hearing loss was strictly conductive in nature. A medical referral was made, but the child has not yet been re-evaluated.

During the pre-evaluation interview, patient #17 reported symptoms similar to those of Menieres disease. She complained of episodic vertigo, severe tinnitus, and sudden loss of hearing. Patient #17 has been treated with Antivert, a non-otoxic drug, for the past five years in order to control attacks of vertigo. The audiologic test results for patient #17 revealed a mild, rising, low frequency sensorineural hearing loss in both ears. This patient was extremely sensitive to loud sounds and minimally cooperative during testing. Although she refused to participate in the entire test battery, her good speech discrimination scores and the absence of significant tone decay on the Olsen-Noffsinger test suggested that the site-of-lesion was primarily in the cochlea.

The other two adult patients with hearing loss, (#18 and #19) showed very similar results. Both patients reported an extensive history of occupational noise exposure and both complained of tinnitus. For the past two years, patient #19 has taken various nonototoxic drugs for a heart condition. Each patient exhibited a moderate/severe high frequency sensorineural hearing loss bilaterally. The absence of tone decay suggested that the site-of-lesion was not retrocochlear, and therefore primarily in the cochlea.

In summary, 79% of the patients tested had normal hearing and 21% had measurable hearing loss. The cochlea was the primary site-of-lesion in three of the four hearing loss cases. The

site-of-lesion for the other hearing loss was in the middle ear.

Since the hearing losses did not follow a pattern, and most were readily identified with specific etiologies, these losses were probably not related to rickets. However, the etiology of the hearing loss of patient #17 could not be accounted for by other factors, therefore, it is possibile that her hearing loss was somehow related to this bone disease.

A compilation of results from the Davies et al. article and this study are shown in Figures 5 to 8. The data in the graphs depict thresholds at 500 and 8000 Hz from both studies as a function of age. Results from the better ear are shown in Figures 5 and 6, for the worse ear in Figures 7 and 8. A more accurate comparison of the studies would have included the PTAs. However, these scores were not published in the Davies et al. article. In general, the patients from the study at CID had better thresholds than the patients in the other study.

Before comparing the two studies, the abnormal thresholds of the three patients (#8, 18, 19) that showed hearing loss most likely due to factors other than rickets, were eliminated from the results. It was stated in the Davies et al. article that the results from patients with a history of noise exposure or ototoxic drugs were not included in the test sample.

The etiology of the hearing loss in patient #17 was unknown.

Therefore, the results for patient #17 were the only abnormal thresholds from the present sample that were plotted on the graphs. All of the remaining thresholds from the CID study were within normal limits (0 to 20 dB HL).

The audiologic findings from the Davies et al. study, however, showed a much larger percentage of abnormal thresholds at both 500 Hz and 8000 Hz. Percentages on each graph of patients with abnormal hearing thresholds are shown in Figure 9.

The results of the two studies were compared to the average expected hearing thresholds according to age as defined by A. Spoor (1967). The 500 Hz and 8000 Hz average thresholds that estimate the amount of hearing loss due to presbycusis, were recorded on the graphs. The purpose of this was to account for any hearing loss that may be attributed to the normal aging process. Since the predicted thresholds for patients with presbycusis are based on averages, it is expected that the normal range of hearing extends 20 dB on either side of the mean.

All of the graphs (Figures 5, 6, 7, and 8) show very similar findings. However, each graph is discussed separately.

Figure 5 includes a comparison of thresholds at 500 Hz in the better ear. All of the thresholds from the CID sample were well within the normal range except two of them. The threshold for the 58 year old patient (#19) was borderline normal and the

threshold for the 40 year old patient (#17) showed a mild loss. The graph showed that 17 of the 18 patients (94.5%) of the thresholds for 500 Hz in the better ear were within the normal range for the CID sample. Therefore, 5.5% of the subjects had a hearing loss. The Davies et al. study showed that five of the 25 subjects, (20%) of the sample tested had abnormal thresholds in the better ear at this frequency.

Thresholds for the better ear at 8000 Hz were plotted in figure 6. None of the thresholds from the CID sample were abnormal, while 13 of the 25 thresholds (52%) from the Davies study were in the abnormal range.

Figure 7 shows hearing thresholds as a function of age in the worse ear at 8000 Hz. Again, all of the thresholds for the patients included in the CID study were within normal limits. Nineteen of the 25 patients (76% of the sample) from the Davies et al. study had abnormal thresholds.

Thresholds for the worse ear of each patient at 500 Hz were plotted on Figure 8. One of the 17 patients, (6% of the CID sample) exhibited an abnormal threshold. In the Davies et al. study, 12 of the 25 patients (48%) showed a hearing loss at 500 Hz in the worse ear.

Each of the figures is very similar. The results showed that the thresholds for a total of 17 out of 18 patients from the CID

study did not exceed the age related hearing thresholds by more than 20 dB. The abnormal thresholds at 500 Hz for the remaining CID patient were only 20 to 25 dB from the mean threshold for subjects the same age. Therefore, there was not an abnormal degradation of hearing sensitivity with age in the present sample.

From these results, it is clear that the hearing sensitivity of patients from the CID study was not affected by rickets.

The results from the Davies et al. sample, however, differ dramatically from the CID data. Most of the thresholds at both frequencies were much worse than those of the CID sample. Also, the Davies et al. study showed a much higher percentage of hearing loss when subjects in the two studies were related by age.

The results also showed that the hearing loss of patients in the Davies et al. sample was frequency dependent. There was a higher percentage of abnormal thresholds at 8000 Hz (76% worse ear; 52% better ear) than at 500 Hz (48% worse ear; 20% better ear).

Since the CID study was a replica of the study conducted by Davies et al. in England, and the patients in both studies had the same type of rickets or osteomalacia, the audiologic results signified that there was some other differing factor between samples that may have caused the hearing loss in the Davies et al. sample.

DISCUSSION

The findings from the CID study do not indicate an association between hypophosphatemic rickets and hearing loss. The combined results of studies do indicate that a hearing loss in children with rickets is very unlikely. However, the effects of this disease on the hearing of adults requires further study because of the discrepancy of results across these studies. Future research should be conducted on a sample of patients 20 years of age or older. Age-appropriate hearing loss predictions for presbycusis should be incorporated.

The occurrence of Menieres disease or endolymphatic hydrops in adults with osteomalacia should also be studied. Results from both studies showed a high incidence of vertigo in the adults. Seven of the 21 adults from the Davies et al. sample and two of the 11 adults from the CID sample reportedly experienced vertigo. Therefore, a total of 9 patients suffered from episodes of vertigo. Three of those patients exhibited symptoms consistent with Menieres disease.

Another area for research may include factors associated with the study of the severity of rickets and degree of hearing loss seen later in life. This is recommended because the test battery and type of rickets in patients across studies were the same, yet the results differed dramatically. One hypothesis is that the differences between samples may be related to the severity

of rickets. Possibly patients in the CID study were only mildly affected by the bone disease. As stated earlier, rickets can be quantified by a bone biopsy.

A second hypotheses is that the differences between samples were related to the treatment of the bone disease. The patients in the present study are receiving medical therapy that helps prevent the abnormal modeling and remodeling of the bones that is seen in patients with rickets.

A third hypothesis is that there may have been some unknown ototoxic effects from treatment used on the patients in the Davies et al. study.

All of these hypothesis may be related to the severity of the bone disease.

SUMMARY

A (1984) study by Davies et al. indicated a higher than expected incidence of hearing loss in patients with X-linked dominant hypophosphatemic osteomalacia or rickets. The present study was conducted to corroborate the previous findings and determine if the patient's age was a contributing factor.

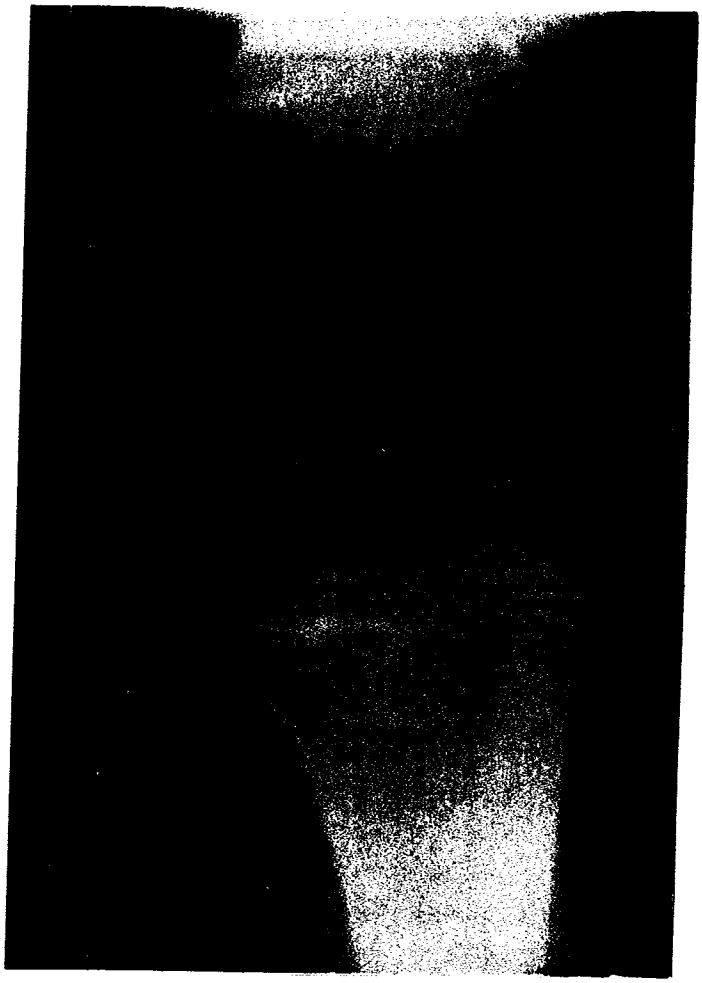
The results of the present study revealed that 79% of the sample exhibited normal hearing sensitivity and only 21% showed measurable hearing loss. Out of the 19 patients with rickets,

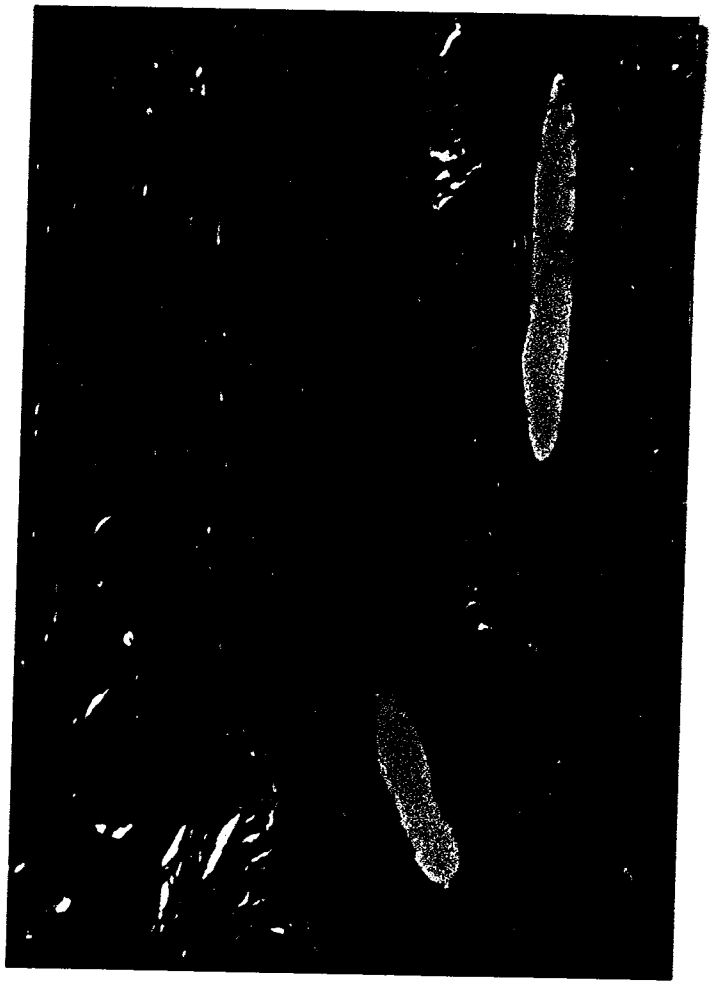
15.5% of the sample had hearing loss that was most likely due to factors not directly associated with bone disease. Only one, or 5.5% of the sample exhibited a hearing loss of unknown etiology, and therefore may be associated with rickets.

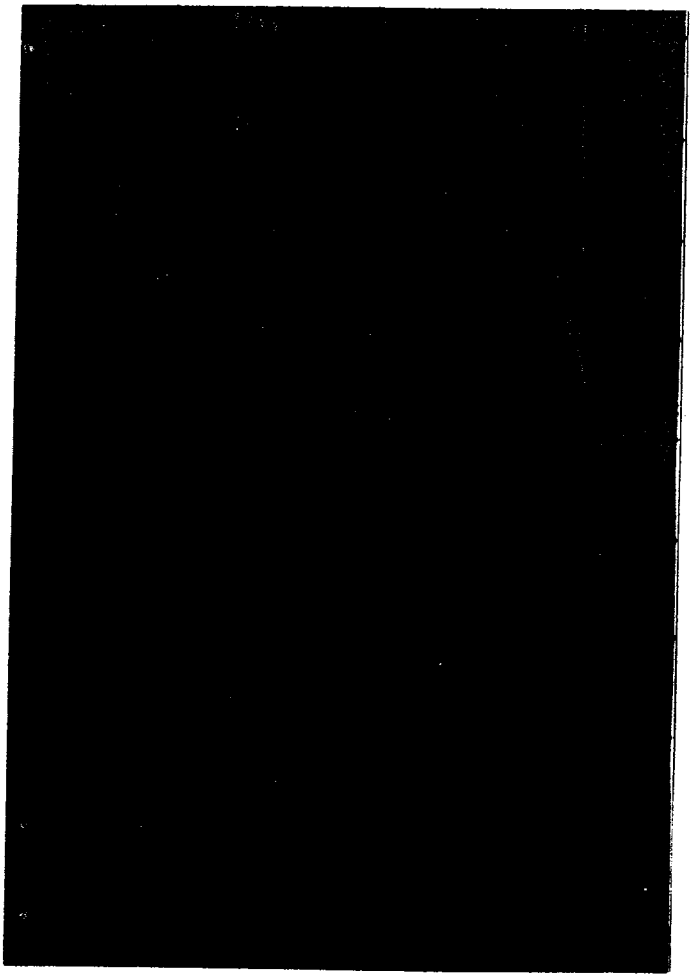
In comparison, the Davies et al. study showed that 76% of the sample had measurable hearing loss that could not be accounted for by other factors, and therefore, was associated with rickets.

The data from both studies did indicate that hearing loss in children with rickets is unlikely, and therefore, if a decrease in hearing sensitivity is associated with rickets, it will probably occur in adults.

Results from the Davies et al. study also suggested that hearing sensitivity was primarily affected in the higher frequency region.







Above left, Fig. 1 An X-ray of the knee joint. The growth plate is identified as a dark area between the bone end and the knuckle. The width of the growth plate and the raggedness of the bone edge would identify rickets in a child.

Above, Fig. 2 A sample of bone biopsy showing well mineralized bone (blue area) and fibrous matrix awaiting mineralization (red area). The percentage of red area seen would identify osteomalacia.

Left, Fig. 3 While the average human reaches the 50th percentile for height, a rickets patient may reach only the 3rd percentile.

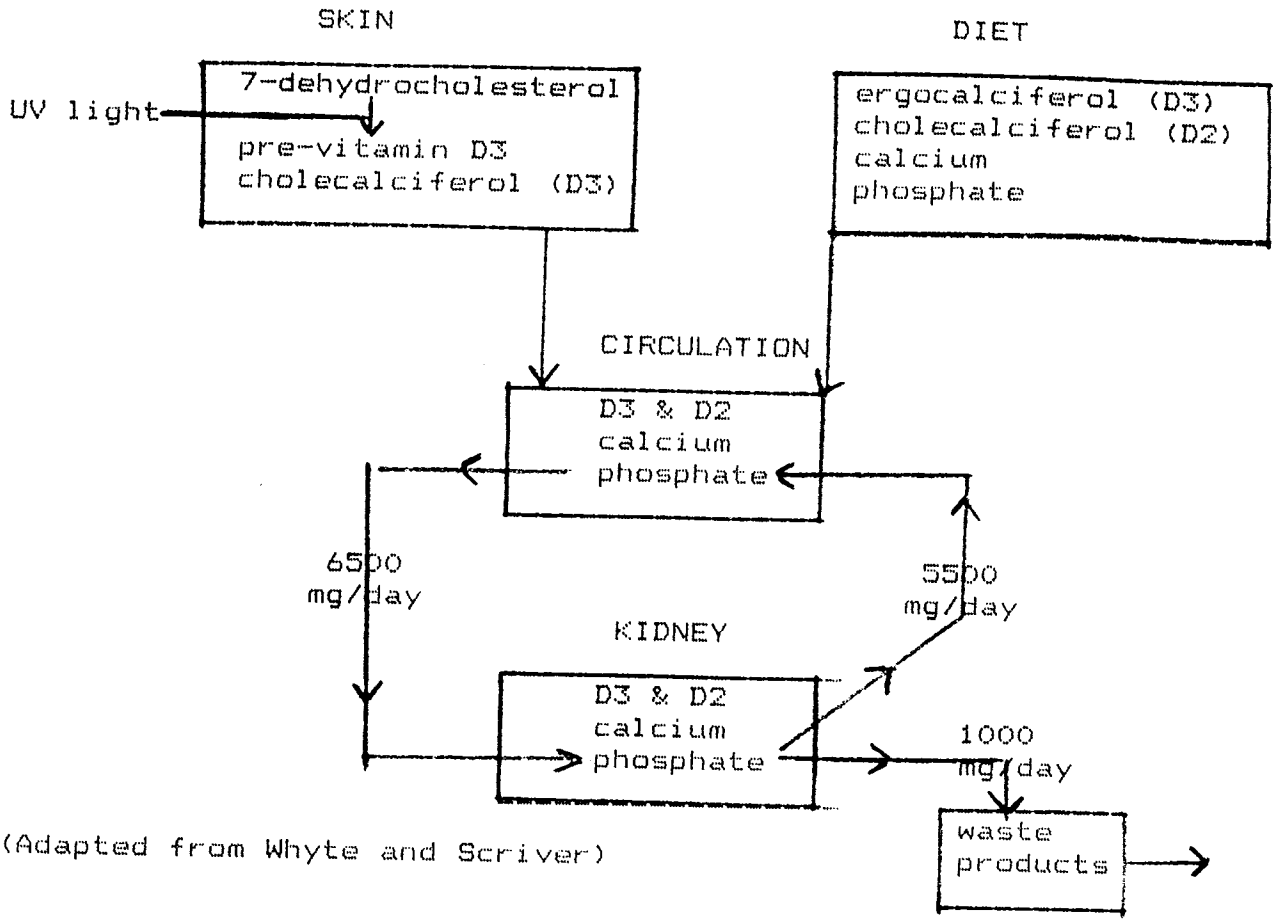


Figure 4. Human cycle of vitamin D, calcium, and phosphate

Ultraviolet light from the sum reacts with a substance in the skin, forming pre-vitamin D3, which is refined into vitamin D3. This is passed along into the circulatory system, or blood stream.

From the foods we eat, we take in vitamin D2, D3, calcium, and phosphate. These substances also enter the blood supply, to be carried throughout the body. It is the combination of vitamins D2 and D3, calcium and phosphate which mineralize the bone, adding a glass-like substance to the fibrous protein in the cells.

After circulation, several times a day, the blood flows through the kidneys, where it is cleansed of impurities. At this time the valuable vitamins and minerals are reclaimed and returned to the circulatory system, while the rest is expelled from the body as waste products.

A normal adult has 6,500 mg of phosphate pass through the kidney daily. On the average, 5,500 mg/day are reclaimed, and put back into the circulatory system. This leaves 1000 mg/day of phosphate which is passed from the body, and this difficet must ve made up through the diet.

In hypophosphatemic rickets, much less is retained, and much more is expelled with the waste materials. When dietary intake of phosphate doesn't maintain equilibrium, rickets can occur.

Table 2

Patient #	Age	Sex	T	V	Ear	HL	PTA dB	SRT HL	Bing	Weber	Impedance IM	Impedance MEP	Config	SDS	TD	ART .5	ART 1	ART 2	ART 4 Hz	Suspected Etyology
1)	1	F	-	-	R	N	25	10	DNT	DNT	.5	-5	peaked	DNT	DNT	DNT				none
					L	N	30	10			.3	-55	peaked							none
2)	4	M	-	-	R	N	10	10	N	Right	.7	-20	peaked	88	none	DNT				none
					L	N	11	5	N		.4	+25	peaked	88	none					none
3)	6	M	-	-	R	N	2	0	N	DNL	.4	-5	peaked	100	DNT	85	85	85	100	none
					L	N	2	0	N		.45	-10	peaked	100		90	85	85	NR	none
4)	8	F	-	-	R	N	3	0	DNT	DNT	.5	0	peaked	DNT	none	DNT				none
					L	N	3	0			.6	0	peaked		none					none
5)	8	F	-	-	R	N	7	5	DNT	DNT	.6	+15	peaked	100	none	DNT				none
					L	N	7	10			.6	+5	peaked	100	none					none
6)	8	F	-	-	R	N	12	5	N	DNL	.5	-100	peaked	98	none	DNT				none
					L	N	7	10	N		.6	-155	peaked	100	none					none
7)	10	F	-	-	R	N	7	5	N	DNL	.9	-100	peaked	92	none	90	85	90	90	none
					L	N	12	5	N		.7	+5	peaked	92	none	85	90	85	NR	none
8)	10	M	-	-	R	N	12	10	N		2.5	0	peaked	100	none	DNT				none
					L	C	25	25	N	L	3.9	-150	peaked	100	none					eustachian tube
9)	13	F	-	-	R	N	10	15	N		.7	0	peaked	100	none	95	90	90	100	none
					L	N	15	10	N	L?	.7	0	peaked	96	none	NR	100	100	110	none
10)	15	F	-	-	R	N	8	5	N	DNL	.2	-25	peaked	100	none	DNT				none
					L	N	7	5	N		.3	-5	peaked	100	none					none
11)	16	F	-	-	R	N	8	5	N	DNL	1.3	+10	peaked	92	none	80	85	85	75	none
					L	N	10	10	P		1.3	+5	peaked	96	none	85	90	85	75	none
12)	28	F	+	-	R	N	5	5	N	DNL	.7	-5	peaked	96	none	100	95	90	95	none
					L	N	7	5	N		.8	0	peaked	96	none	95	90	90	90	none
13)	33	F	-	-	R	N	8	5	N	DNL	.6	-15	peaked	96	none	DNT				none
					L	N	11	0	N		.7	-20	peaked	100	none					none
14)	35	F	+	+	R	N	8	10	N	DNL	.6	-5	peaked	96	none	DNT				none
					L	N	10	15	N		.5	-10	peaked	100	none					none
15)	37	F	-	-	R	N	15	15	N	DNL	.8	+10	peaked	96	none	DNT				none
					L	N	10	10	N		.9	+15	peaked	100	none					none
16)	39	F	+	+	R	N	8	5	N	DNL	.5	-10	peaked	96	none	90	90	85	85	none
					L	N	5	0	N		.5	-10	peaked	100	none	90	90	90	85	none
17)	40	F	+	+	R	SN	20	25	DNT	DNT	refused			96	none	refused				unknown
					L	SN	22	25			refused			92	none	refused				unknown
18)	43	M	+	-	R	SN	12	15	N		.7	-10	peaked	88	none	70	95	100	NR	noise exposure
					L	SN	18	20	N	L	.8	-15	peaked	92	none	CNT				noise exposure
19)	58	M	+	-	R	SN	17	25	N	DNL	.3	-50	peaked	90	none	DNT				noise exposure
					L	SN	18	20	N		.5	-55	peaked	92	none	DNT				noise exposure

T=Tinnitus V=Vertigo +=positive -=negative
HL=Hearing loss, N=none, C=concuctive, SN=sensorineural
Bing, N=negative, P=positive for conductive component
Weber, DNL=did not lateralize DNT=did not test
IM=immittance @ 220 Hz in EV, MEP=middle ear pressure in da Pa
config=configuration

SDS=speech discrimination score in % correct
TD=tone decay
ART=Acoustic Reflex Threshold in dB HL re stimulus ear

Fig. 5

BETTER EAR at 500 Hz

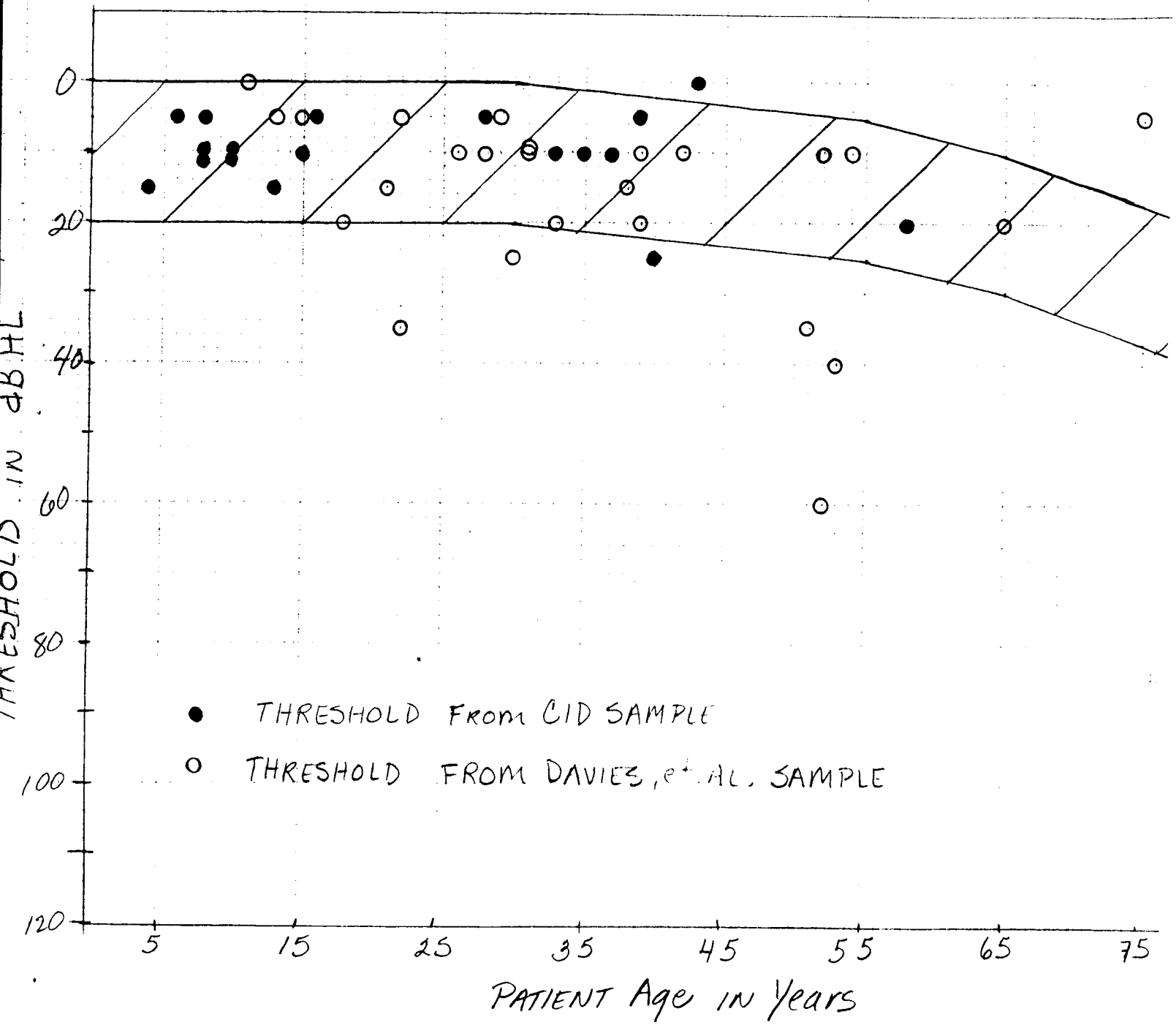


Fig. 6

BETTER EAR 8000 Hz

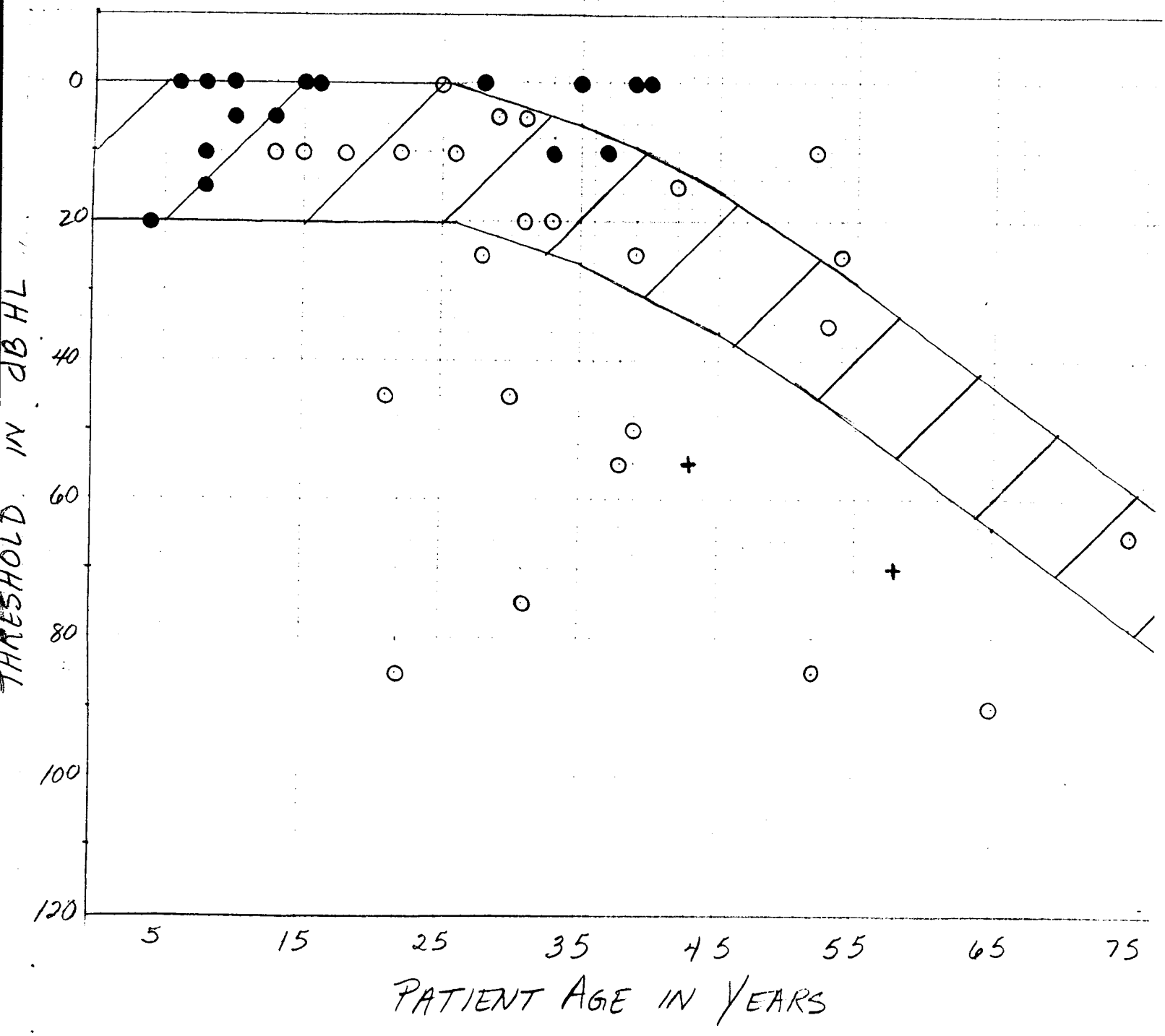


Fig. 7
Worse Ear at 8000 Hz

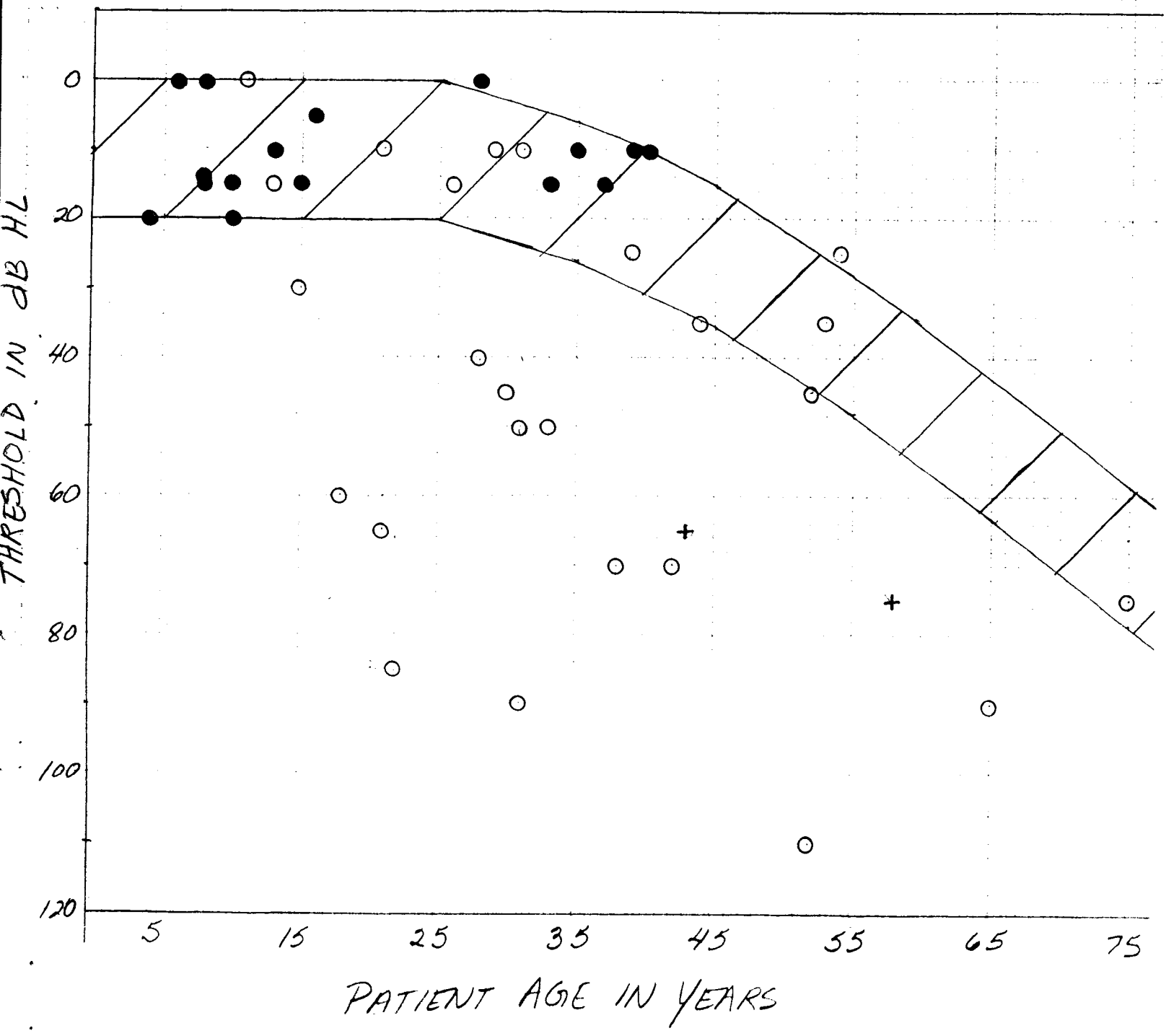


Fig. 8

WORSE EAR at 500 Hz

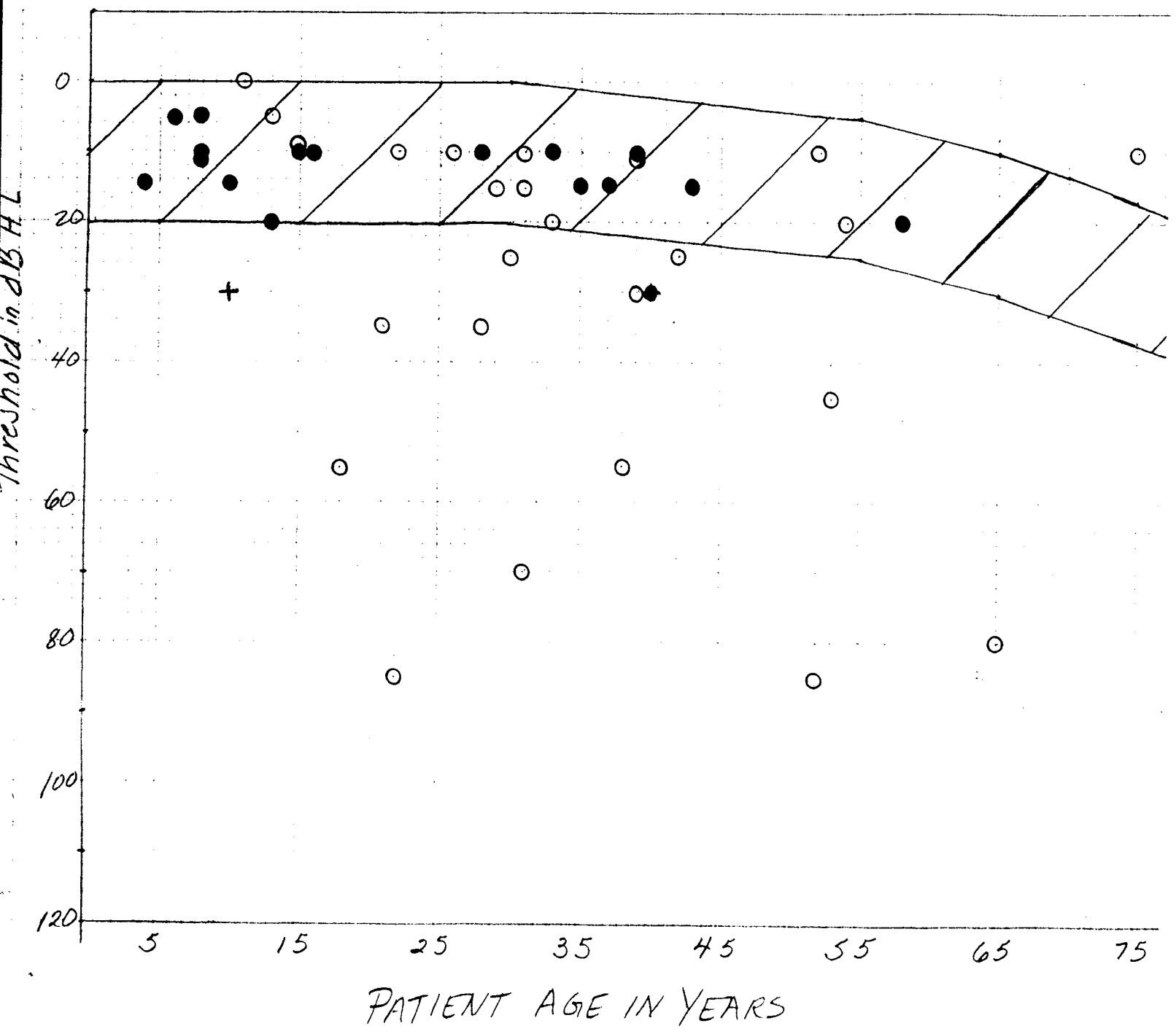
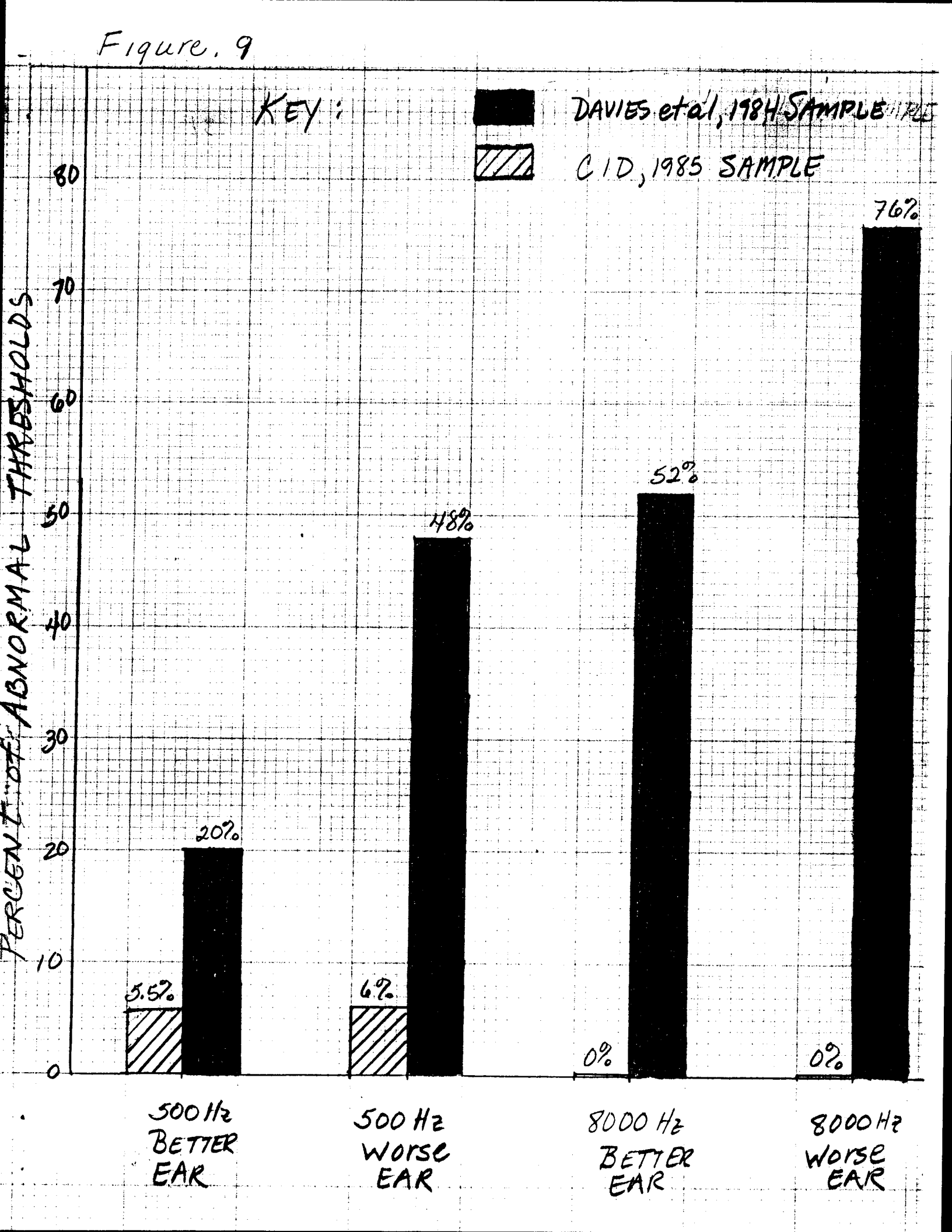


Figure. 9


KEY:
DAVIES et al, 1984 SAMPLE
CID, 1985 SAMPLE
PERCENT OF ABNORMAL THRESHOLDS
0
10
20
30
40
50
60
70
80
5.5%
20%
6%
48%
0%
52%
0%
76%
500 Hz BETTER EAR
500 Hz WORSE EAR
8000 Hz BETTER EAR
8000 Hz WORSE EAR

BIBLIOGRAPHY

Brookes, G. B. (1983). Vitamin D deficiency--a new cause of cochlear deafness. The Journal of Laryngology and Otology, 97, 405-420.

Davies, M. (1984). Impaired hearing in X-linked hypophosphatemic (vitamin-D-resistant) osteomalacia. Annals of Internal Medicine, 100, 230-232.

Scriver, C.R., & Tenenhouse, H.S. (1981). On the heritability of rickets, a common disease (Mendel, mammals, and phosphate). The Johns Hopkins Medical Journal, 149, 179-187.

Stamp, T. C. B., & Baker, L. R. I. (1976). Recessive hypophosphataemic rickets, and possible aetiology of the "vitamin-D-resistant" syndrome. Archives of Disease in Childhood, 51, 360-365.

Spoor, A. (1967). Presbycusis values in relation to noise induced hearing loss. Int. Audiology, 6, 48-57.

Weir, N. (1977). Sensorineural deafness associated with recessive hypophosphataemic rickets. Journal of Laryngology and Otology, 91,717-722.

Whyte, M.P. (1985) Rickets and osteomalacia. In R.E. Rakel (Ed.), Conn's Current Therapy (pp. 452-462). Philadelphia: W.B. Saunders Company